

Microlymphoma, A Rare Clinicopathologic Entity During the Evolution of an Aggressive Lymphoma: A Case Report

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Microlymphoma, A Rare Clinicopathologic Entity During the Evolution of an Aggressive Lymphoma: A Case Report.

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Introduction:

We report a patient case presenting with generalized lymphadenopathy on work-up diagnosed with HHV-8 associated Multicentric Castleman Disease (HHV-8+MCD) with microlymphoma in the setting of newly diagnosed HIV. Microlymphoma is a very rarely reported clinicopathological entity, which is described as a precursor to aggressive plasmablastic lymphoma (PBL), mostly seen in HIV patients.

Case:

A 57-year-old male presented to the ED with cough, SOB, intermittent fevers and generalized weakness since three weeks. Initial workup showed severe pancytopenia, normal blood chemistry, elevated CRP 112 mg/L and peripheral smear showing plasmacytoid and atypical lymphocytes. An HIV assay was positive with decreased CD4/CD8 ratio. Bone marrow biopsy showed hypercellularity, but no signs of lymphoproliferative neoplasm. CT scan demonstrated generalized lymphadenopathy with splenomegaly. The lymph node biopsy showed an overall preserved architecture with open sinuses and numerous interfollicular small plasma cells. In focal areas, follicles with concentric layering of plasmablasts within the mantle zone reminiscent of Castleman disease were seen. These follicles were more expanded in other areas with confluent sheets of plasmablasts (Fig. 1). Immunohistochemistry demonstrated the internodular small plasma cells to be CD138+ with polytypic kappa and lambda light chain expression. The clusters and nodules of plasmablasts were lambda light chain restricted and HHV-8+ by in-situ hybridization (Fig. 2). The patient was initiated on HAART therapy with rituximab. After 4-weeks of treatment, he was seen in clinic with improved symptoms and normal counts.

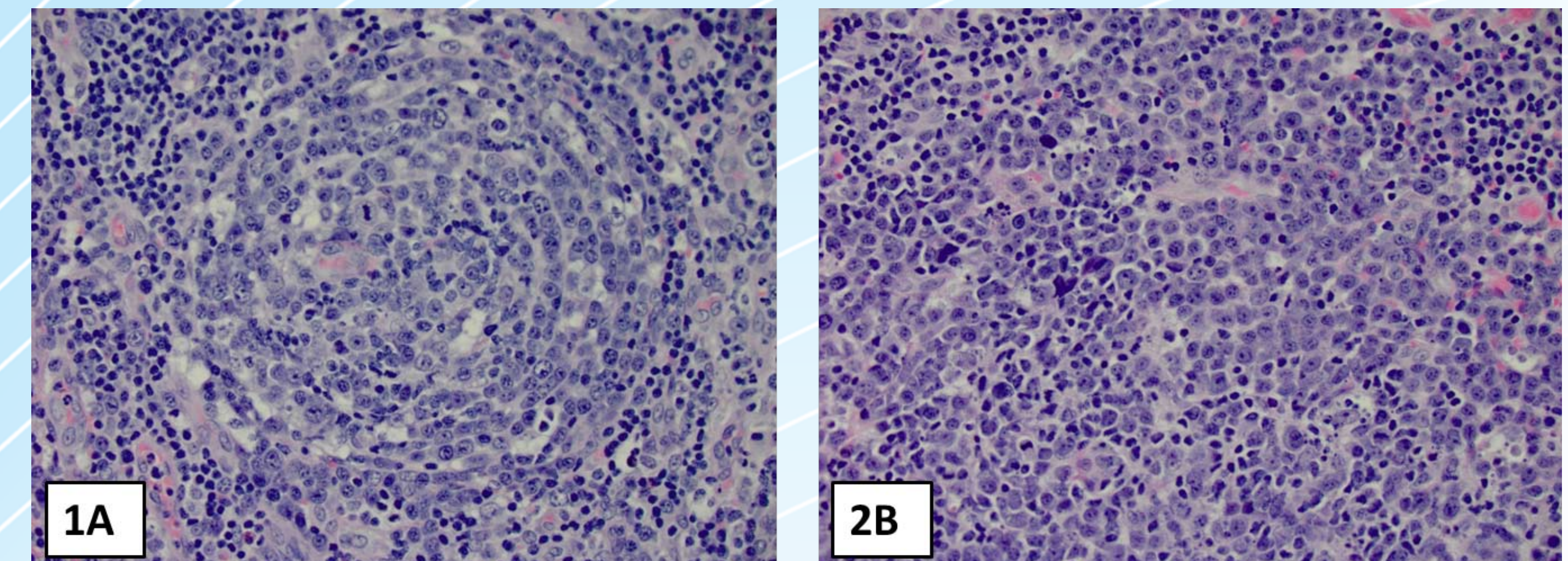


Figure 1: A. B-cell follicle with expanded mantle zone infiltrated by numerous large plasmablastic cells with dense amphophilic cytoplasm and nuclei with one or two prominent nucleoli. B. Large follicle with confluent sheets of plasmablast [hematoxylin-eosin, original magnifications X400].

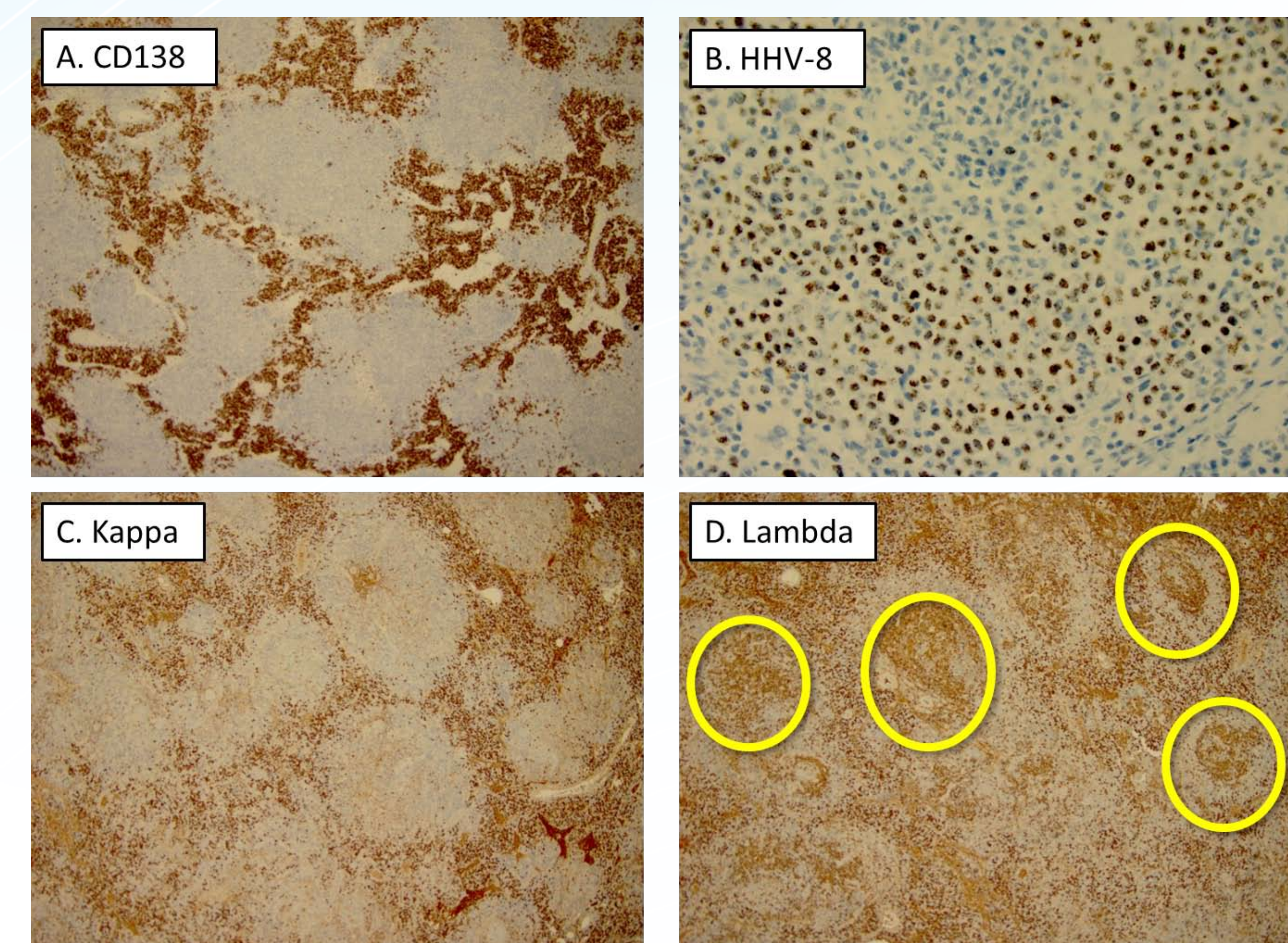


Figure 2: Immunohistochemistry shows CD138 -positive plasma cells (A) with subsets expressing polytypic kappa and lambda light chain located in the internodular region (C & D). Plasmablasts are lambda light chain restricted and are confined to the centers of the follicles (blue circles) (D). Plasmablasts are positive for HHV-8 stain by in-situ Hybridization (B) original magnifications X200 [A, B, C and D].

Discussion:

Microlymphoma is a rare intermediate clinicopathological entity that is described during the evolution from HHV-8+MCD to the onset of an aggressive PBL. Albeit having no frank histopathologic evidence of aggressive lymphoma in our patient, the clinical and histological features were compatible with HHV-8+MCD with confluent clusters of HHV-8+ plasmablasts coalescing to form microlymphomas. Given the rarity of PBL and the poor prognosis associated with HIV/HHV-8+ cases, understanding of the natural history of PBL is important to initiate early treatment.

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